CONGENITAL HEART DISEASE

Transcatheter closure of ventriculopulmonary artery communications in staged Fontan procedures

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Heart 2007;**93**:510–513. doi: 10.1136/hrt.2006.093757

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Accepted 24 October 2006
Published Online First 12 December 2006

Background: Ventricle-pulmonary artery connections in patients after the Fontan procedure lead to ineffective volume loading and can cause long term problems. In patients with a cavopulmonary shunt anterograde pulmonary blood flow is frequently maintained, but can cause significant volume loading of the heart or complicate the subsequent Fontan procedure.
Objective: To evaluate the use of transcatheter closure of a ventricle-pulmonary artery communication in the setting of a cavopulmonary shunt or after the Fontan procedure.
Patients and methods: Retrospective study at a tertiary referral centre. Eight patients (age 1.5–18 years, mean 7.8 years). Indications: cardiac failure or persistent pleural effusions after cavopulmonary shunt (n = 2) or after Fontan (n = 3) and abolishing the volume load of the single ventricle prior to Fontan completion (n = 3).
Results: Devices used: Rashkind Umbrella (n = 1), Amplatzer PDA (n = 7) and Amplatzer ASD (n = 1). One patient required two devices. There were no procedural complications. All 3 patients with prolonged pleural effusions (1 post CP shunt and 2 post Fontan) showed complete resolution between 4 and 10 days after catheter closure. Two patients underwent transcatheter occlusion for progressive ventricular dilatation and cardiac failure. The first patient was post Fontan and showed gradual improvement in ventricular function. The second patient (post CP shunt) was in end stage cardiac failure due to severe AV valve regurgitation. The patient died 48 hours after an uncomplicated procedure due to ventricular failure and electromechanical dissociation (non-procedure-related cardiac death). Three patients underwent catheter closure to off-load the systemic ventricle prior to the Fontan procedure. The device had to be removed prior to release in one patient, due to unsatisfactory position.
Conclusions: Transcatheter closure of ventricle-pulmonary artery communication is a safe and effective technique in the treatment of selected patients after cavopulmonary shunt or Fontan procedure with early or late complications due to inappropriate pulmonary blood flow. This intervention should also be considered in the preparation for the Fontan procedure in selected patients with ventricular overload.

The management of patients with complex congenital heart disease that are not amenable to complete, biventricular repair generally involves a series of palliative operations, which culminate in the Fontan procedure.[1] The Fontan procedure is most commonly performed as a staged procedure. This involves the formation of a superior bidirectional cavopulmonary (CP) shunt during infancy and then rerouting of the inferior caval venous return to the pulmonary arteries at a later operative procedure. A number of centres have advocated that restrictive antegrade pulmonary blood flow via a stenotic pulmonary outflow tract should be preserved during the creation of a superior CP shunt.[2–4] At the time of the Fontan procedure, it is necessary to abolish all sources of additional pulmonary blood flow, including antegrade pulmonary blood flow, to prevent obligate volume loading of the heart. However, this will result in an acute reduction in the ventricular preload, which may cause diastolic dysfunction in the early post-operative period.[5] In cases in whom forward flow from the ventricle to the pulmonary arteries persists after the Fontan procedure, there is a risk of persistent pleural effusions or progressive ventricular failure.

This report describes the technique and results of interventional catheter closure of ventriculopulmonary arterial communications in the setting of a superior CP shunt or after the Fontan procedure.

PATIENTS

Between January 1988 and July 2004, 406 patients underwent the Fontan procedure at Birmingham Children's Hospital, Birmingham, UK at a median age of 4.7 years (range, 9 months to 52 years). Acute Fontan takedown was required in 2.2% (n = 9). In-patient mortality was 3% (n = 12). Early survival with a Fontan circulation was 94.8% (n = 385). Actuarial survival following the Fontan procedure was 92.7% (1.3%) at 1 year, 90.2% (1.7%) at 5 years and 86.1% (2.3%) at 10 years, postoperatively. The Fontan procedure was a staged procedure in 285 (70%) patients.

Ventriculopulmonary arterial communications after superior CP shunt or completed Fontan procedure were closed by interventional cardiac catheterisation techniques in eight patients who represent the study population (table 1).

Indications for closure were, firstly, cardiac failure or persistent pleural effusions after superior CP shunt (n = 2) or after Fontan procedure (n = 3) or, secondly, abolishing the volume load of the systemic ventricle prior to completion of the Fontan procedure (n = 3).

Patients were aged from 1.5 to 18 years at the time of intervention (mean 7.8 years). There were four males and four females.

METHODS

All procedures were performed under general anaesthesia, and endotracheal intubation and ventilation. In patients, a right internal jugular venous approach was used after superior CP shunt. This was complemented by either femoral

Abbreviations: AV, atrioventricular; CP, cavopulmonary; PDA, patent ductus arteriosus

Table 1 Patient and procedural data

No	Sex	Diagnosis	Timing of procedure/status	Time interval since surgery	Indication	Type of device	Procedural outcome
1	M	DILV, TGA, PS/sub PS	Post-Fontan	2 weeks	Prolonged bilateral pleural effusions	17 mm Rashkind double umbrella	Resolution of effusions
2	F	Unbalanced AVSD, DORV, PS	Post-Fontan	6 years	Progressive ventricular dilation and failure	PDA Amplatz 8/6	Improvement in ventricular function and exercise tolerance
3	M	RAI, unbalanced AVSD, PS/sub PS	Post-Fontan	3 weeks	Prolonged bilateral chylous effusions	PDA Amplatz 8/6 and Amplatz ASD	Resolution of effusions
4	M	DILV, TGA, sub PS	Pre-Fontan	–	Off loading ventricle preparation for Fontan	PDA Amplatz 10/8	Successful completion of Fontan after 6 weeks
5	F	Mitral atresia, DORV, TGA, PS	Pre-Fontan	–	Off loading ventricle preparation for Fontan	PDA Amplatz 8/6	Device not in satisfactory position—removed
6	F	Dextrocardia, AVSD, DORV, TGA, PS	Post-CP shunt	12 months	Severe AVVR, end-stage ventricular failure	PDA Amplatz 10/8	Died 48 h post catheter due to end-stage ventricular failure
7	M	DILV, TGA, sub PS, congenital CHB	Post-CP shunt	3 months	Prolonged chylous pleural effusions	PDA Amplatz 10/8	Resolution of effusions
8	F	Mitral atresia, DORV, TAPVD, CoA	Pre-Fontan	–	Off loading ventricle preparation for Fontan	PDA Amplatz 6/4	Successful completion of Fontan after 3.5 months

ASD, atrial septal defect; AVSD, atrioventricular septal defect; AVVR, atrioventricular valvar regurgitation; CHB, complete heart block; CoA, coarctation of aorta; DILV, double inlet left ventricle; DORV, double outlet right ventricle; F, female; M, male; PDA, patent ductus arteriosus; PS, pulmonary stenosis; RAI, right atrial isomerism; sub PS, sub pulmonary stenosis; TAPVD, total anomalous pulmonary venous drainage; TGA, transposition of great arteries.

venous or arterial approaches. All patients received Heparin 50 U/kg, and complete haemodynamic and angiographic studies were performed. Morphological assessment of the ventriculopulmonary arterial communications was completed by both transthoracic and transoesophageal ultrasound studies.

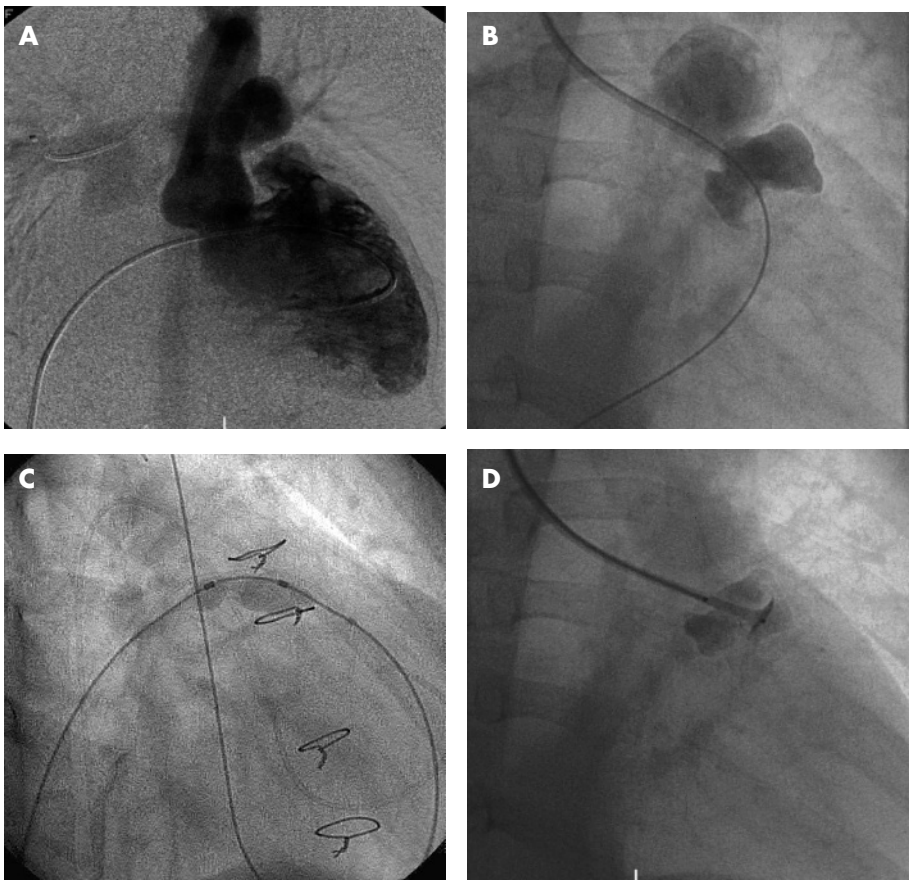

Figure 1 (A) Ventriculogram showing antegrade ventriculopulmonary artery communication. (B) Site of pulmonary artery band. (C) Balloon sizing for device selection. (D) Patent ductus arteriosus Amplatzer device prior to release.

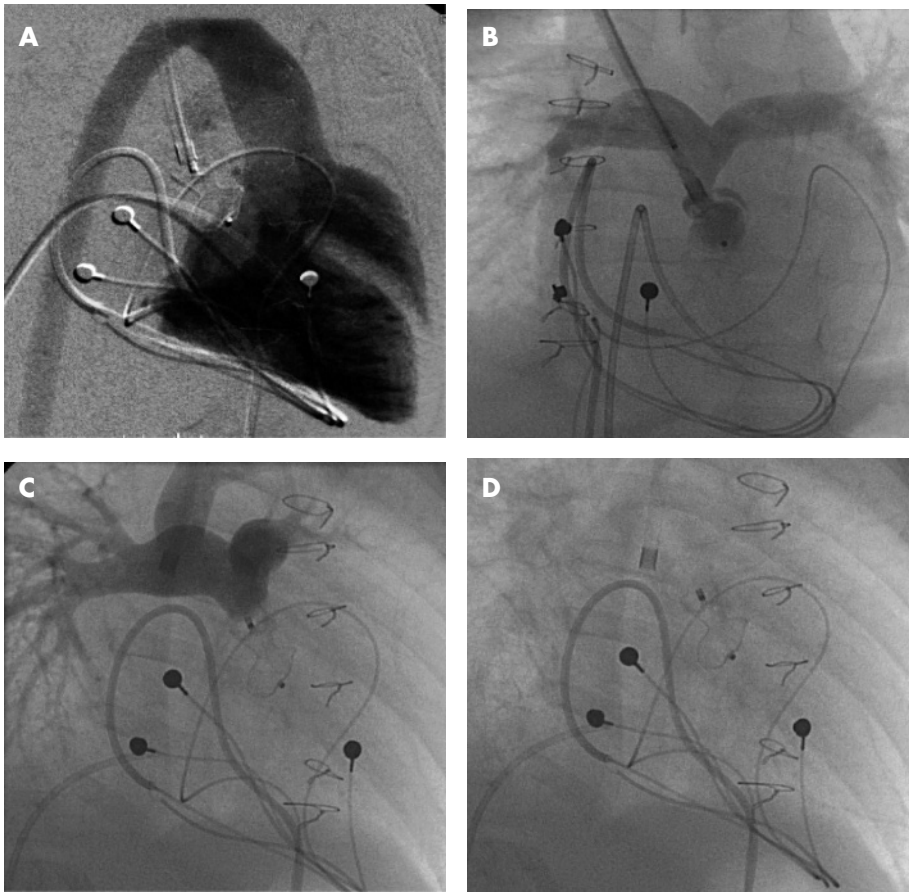

Figure 2 (A) Ventriculogram with device deployment. (B) Patent ductus arteriosus Amplatzer device occluding ventriculopulmonary artery communication. (C) Angiogram of superior vena cava to assess pulmonary arteries after device release. (D) Screening device after release.

After placement of a guidewire across the ventriculopulmonary arterial communication, a standard balloon valvuloplasty catheter was placed across the lesion and gently inflated to obtain an imprint of the narrowest point of the stenosis, and to reassess haemodynamics and atrioventricular (AV) valve function during temporary occlusion. Measurements of the narrowest diameter of the balloon were obtained. In the cases who underwent closure using an Amplatzer patent ductus arteriosus (PDA) device (AGA Medical Corporation, Golden Valley, Minnesota, USA), we selected a device with a diameter 2–3 mm greater than the minimal balloon diameter.

In the first patient, a 17 mm Rashkind double-umbrella device delivered via an 11 French Mullins sheath was used. In six patients, an Amplatzer PDA occluder was delivered via the appropriate-sized delivery sheath. One patient underwent catheter closure using Amplatzer atrial septal defect and PDA devices (one each).

STATISTICAL ANALYSIS

Haemodynamic parameters were assessed before and after occlusion. Differences were analysed using a paired t test, $p<0.05$ was considered significant.

RESULTS

Temporary test occlusion of the antegrade pulmonary blood flow decreased the mean (range) pulmonary artery pressure from 17.2 (13–21) to 14 (11–18) mm Hg ($p<0.05$). The mean (range) transpulmonary gradient decreased from 8 (4–11) to 5.5 (4–7) mm Hg ($p<0.05$). Mean (range) systemic ventricular end-diastolic pressure decreased from 10.8 (10–13) to 9.5 (8–11) mm Hg ($p<0.05$). Mean (range) systemic oxygen saturation in patients 4–8 after CP shunt decreased from 81.8% (78–88%) to 75% (72–78%) ($p<0.05$). Two patients had immediate reduction in AV valve regurgitation during occlusion. There was no increase in AV valve regurgitation in any of the eight patients.

Three patients had prolonged pleural effusions, one 12 weeks after a CP shunt (1.5 years) and the other two 2 and 3 weeks after Fontan procedure (7 and 10 years), in whom the antegrade pulmonary blood flow could not be abolished at the time of surgery due to dense adhesions around the main pulmonary artery. In all three patients there was rapid complete resolution of pleural effusions following occlusion of the ventriculopulmonary artery communication.

Two patients presented with progressive ventricular dilatation and cardiac failure due to antegrade pulmonary blood flow. The first patient presented at 18 years of age, 6 years after Fontan procedure, with marked ventricular volume load and dyspnoea. She showed signs of gradual improvement in ventricular function and effort tolerance after the catheter occlusion of the ventriculopulmonary artery communication. The second patient, 1.5 years old, presented with severe AV valve regurgitation and end-stage cardiac failure 10 months after CP shunt. Catheter intervention was performed as a salvage to reduce excessive volume loading on the ventricle. The procedure was successful and uneventful. The patient returned to the intensive care unit in a stable condition in sinus rhythm, with normal AV conduction and no increase in the degree of AV valvar regurgitation. However, the patient died 48 h after the procedure due to ventricular failure and electromechanical dissociation. Postmortem examination was declined by the family.

Prior to Fontan procedure, three patients underwent transcatheter occlusion of ventriculopulmonary artery communication in an attempt to reduce the volume load on the systemic ventricle and facilitate completion of the Fontan procedure. In

one patient (6 years), with previous banding of the main pulmonary artery, the PDA Amplatz device had to be removed prior to release, as this was protruding within the lumen of the central pulmonary artery. She subsequently underwent successful completion of the Fontan procedure with surgical ligation of main pulmonary artery. The second patient (6 years) had reduction in transpulmonary gradient and ventricular dilatation followed by successful completion of the Fontan procedure within 6 weeks. The third patient (12 years), who had previously failed a Fontan procedure, requiring takedown, had successful completion of the Fontan procedure within 4 months after the transcatheter occlusion.

DISCUSSION

The Fontan procedure redirects the systemic venous return to the pulmonary arteries, and the pulmonary venous return to the systemic circulation. Residual forward flow from the ventricle to the pulmonary artery, via either a native pulmonary outflow tract or a previously banded or ligated main pulmonary artery, leads to unnecessary and ineffective pulmonary blood flow and ventricular volume overload. This in turn can lead to persistent pleural effusions or ventricular failure. Treatment of such lesions has only been rarely reported in the literature.[6–8]

In patients with functionally univentricular hearts and restrictive antegrade pulmonary blood flow, this is often maintained during the initial CP shunt procedure. However, in some cases, this can lead to excessive ventricular volume load and result in ventricular failure.[9 10]

This report describes our experience with transcatheter occlusion of such lesions in a variety of patients with symptoms, both after CP shunt and after completed Fontan procedure. The morphology of the residual ventriculopulmonary arterial communication was assessed using a combination of ultrasound, angiographic and balloon-sizing techniques (fig 1). The immediate effects of occlusion on pulmonary artery pressures and systemic saturations were assessed by temporary balloon occlusion before proceeding to permanent transcatheter occlusion. There was a significant decrease in the pulmonary arterial pressure, systemic ventricle end-diastolic pressure and transpulmonary gradient. There was also a significant decrease in arterial oxygen saturation in the subgroup of patients after CP shunt, but this was well tolerated. The most suitable device for occlusion was the Amplatzer PDA device, since its introduction in 1998. In this series, we have not experienced any cases of conduction abnormalities or AV valve dysfunction related to the procedure (fig 2).

Following our initial results and success with this technique, we have expanded its use to the catheter preparation for the Fontan procedure in selected patients with CP shunt and antegrade pulmonary blood flow. This is to offload the systemic ventricle prior to surgery and to reduce the early postoperative diastolic dysfunction commonly seen in such patients.[5] In particular, in patients with transposed great arteries, in whom surgical dissection of the main pulmonary artery for ligation during the Fontan procedure may be difficult or hazardous, such an approach is judged to reduce the length and complexity of the Fontan procedure.

CONCLUSIONS

Transcatheter closure of ventriculopulmonary artery communication is a safe and effective technique in the treatment of selected patients after CP shunt or Fontan procedure with early or late complications due to inappropriate pulmonary blood flow. This intervention should also be considered prior to the Fontan procedure in selected patients with systemic ventricular overload.

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Competing interests: None declared.

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